CASE REPORT

Child with aplastic anemia: Anesthetic management

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ABSTRACT

Aplastic anemia is a rare heterogeneous disorder of hematopoietic stem cells causing pancytopenia and marrow hypoplasia with the depletion of all types of blood cells. This results in anemia, neutropenia and thrombocytopenia, which pose a challenge to both surgical and anesthetic management of such cases. We report a child with aplastic anemia who sustained traumatic ulcer on the arm and underwent split-thickness skin grafting under general anesthesia. There are only two case reports on anesthetic considerations in aplastic anemia patients in the literature. The anesthetic management is challenging because of the rarity of the disease, associated pancytopenia and immunosuppression.

Key words: *Aplastic anemia, pancytopenia, anesthesia*

INTRODUCTION

Aplastic anemia is a rare hemopoietic stem cell disorder that results in pancytopenia and hypocellular bone marrow arising from a variety of disease states, including acquired aplastic anemia and diverse congenital marrow failure syndromes.[1] In acquired aplastic anemia, there is immune-mediated destruction of hemopoietic stem cells triggered by environmental exposures, such as to drugs, viruses and toxins, resulting in anemia, thrombocytopenia and neutropenia. Hence, patients with aplastic anemia most commonly present with symptoms of anemia, skin or mucosal or retinal hemorrhage and, rarely, infections.[2] These risks are increased several folds in the perioperative period, and any surgical intervention in these patients should be undertaken only after preoperative optimization. We report a child with aplastic anemia who sustained traumatic ulcer on the arm and underwent split-thickness skin grafting (STSG) under general anesthesia. There are only two case reports on anesthetic considerations in aplastic anemia patients in the literature.[3,4] The anesthetic management is challenging because of the rarity of the disease, associated pancytopenia and immunosuppression.

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CASE REPORT

A 7-year-old male (22 kg) student, known case of aplastic anemia for 18 months, was admitted with traumatic ulcer over the right arm after 7 days following fall from chair. The child had high-grade fever for the last 3 days, for which he was managed conservatively with intravenous antibiotics, fresh packed red cells and platelet transfusions. The large nonhealing ulcer in the right arm being the focus of infection, the child was considered for STSG of the wound.

The child was diagnosed severe aplastic anemia (SAA) 18m back with bone marrow cellularity less than 5% and neutrophils 0.3×10^9/L. Because he did not have an human leukocyte antigen (HLA)-matched sibling, he was given immunosuppressive therapy 1 year prior to admission with antithymocyte globulin (ATG) and cyclosporine, but he did not respond to it and had multiple admissions for fever and bleeding episodes. He had no history of congenital skin or skeletal anomalies, drug exposure, loss of consciousness, seizures, jaundice and joint pains.

On admission, blood investigations showed leukocytopenia (white blood cells 1.24×10^9/L), anemia (hemoglobin 6.7 g/dL) and thrombocytopenia (platelets 12×10^9/L). His preoperative optimization was performed in collaboration with the hematology department. He received 4 units of

irradiated fresh packed red cells and his hemoglobin increased to 8.8 gm/dL and white blood cells count to 1.45 × 10^9/L. In view of the short half-life of the platelets, immediately preoperatively, 2 units of single donor plasma (SDP) were transfused, aiming platelets counts above 50 × 10^9/L. Normal chest roengtogram, liver function tests and viral markers ruled out any chest infection and antecedent hepatitis. Preoperative preparation to prevent bleeding and infection included good orodental hygiene, antiseptic mouthwash, prophylactic antibiotics and antifungals and nursing in isolation.

He underwent STSG under general anesthesia with monitoring, which included a continuous electrocardiogram, noninvasive blood pressure, oxygen saturation, end-tidal carbon dioxide, temperature and urine output. Anesthetic induction was performed with intravenous fentanyl, propofol and atracuruim after 3 min of preoxygenation and a ProSeal laryngeal mask airway (LMA) (No. 2.5) was inserted under strict asepsis. Both the surgical and the anesthesia team followed strict asepsis during the surgery as well as postoperatively. The surgery was uneventful and lasted for 80 min, with blood loss of approximately 100 mL being replaced by packed red cells. Eight units of platelets were transfused intraoperatively. Extubation was smooth and intravenous tramadol was given for postoperative pain relief. Regular blood counts were performed postperatively. He did not have any postprocedural complications. On the 7th postoperative day, he had 30% of skin engraftment, and the donor site of the left thigh had also healed slowly.

DISCUSSION

Aplastic anemia is a rare heterogeneous disorder of hematopoietic stem cells causing pancytopenia and marrow hypoplasia with the depletion of all types of blood cells.[3] This results in anemia, neutropenia and thrombocytopenia, which pose a challenge to both surgical and anesthetic management of such cases. To define aplastic anemia, there must be at least two of the following: (i) hemoglobin <10 g/dL, (ii) platelet count <50 × 10^9/L or (iii) neutrophil count <1.5 × 10^9/L, which were satisfied in the patient. The severity of the disease is graded according to the blood count parameters and bone marrow findings as summarised in Table 1,[2] which indicated that the child had SAA. Regardless of the severity of aplastic anemia, pancytopenia itself poses an increased surgical risk.

The incidence of aplastic anemia is two to three-times higher in East Asia than in Europe and North America.[2] To avoid perioperative complications, preoperative optimization is essential prior to any elective surgery. A multidisciplinary team approach that includes an experienced hematologist and a well-equipped blood bank is recommended to formulate an appropriate management plan for the patient.

Table 1: Definition of severity of aplastic anemia

Severe AA	BM cellularity <25%, or 25–50% with <30% residual hemopoietic cells*
	2/3 of the following:
	Neutrophil count <0.5 × 10^9/L
	Platelet count <20 × 10^9/L
	Reticulocyte count <20 × 10^9/L
Very severe AA	As for severe AA but neutrophils <0.2 × 10^9/L
Nonsevere AA	Patients not fulfilling the criteria for severe or very severe aplastic anemia

*Cellularity should be determined by comparison with normal controls

Absence of skin, nail and skeletal abnormalities in our patient ruled out any congenital form of aplastic anemia, i.e. fanconi anemia, dyskeratosis congenita, Shwachman-Diamond syndrome and amegakaryocytic thrombocytopenia.[2] A careful drug and occupational exposure history should be taken and any putative drug should be discontinued and should not be given again to the patient.[2]

Blood pressure and renal and liver function tests should also be monitored regularly while on cyclosporine and fever, rigors, rash, hypertension or hypotension and fluid retention should be watched for while on ATG.

Bleeding and infection are the dreaded complications that must be prevented. Patients may present for surgery with anemia and thrombocytopenia to a degree that transfusions are necessary. Red cell and platelet transfusion support is essential for patients with aplastic anemia to maintain a safe hemoglobin level (>8 g/dL in the absence of comorbidities and 10 g/dL in patients with cardiac disease).[5] Platelet transfusions must be given in the immediate preoperative period (short half-life) to achieve appropriate levels as recommended by the BCSH guidelines, and a preprocedure platelet count checked to ensure that the level has been achieved. Patients who become refractory to platelet transfusions should be screened for HLA antibodies after excluding causes of platelet refractoriness, such as infection and drugs.[2] Other important measures to help prevent bleeding include good dental hygiene, the use of oral tranexamic acid, use of fresh blood products perioperatively and meticulous surgical hemostasis.

General anesthesia is the only technique available because thrombocytopenia contraindicates use of regional anesthesia. We used LMA to avoid any laryngoscopy-induced trauma. Both induction and emergence should be smooth and atraumatic, as was ensured in our case.

Another important aspect is prevention of infection in such patients. The risk of infection is determined by the patient's neutrophil and monocyte counts.[2] It is mandatory

to follow strict aseptic precautions for all anesthetic and surgical manoeuvres, like intravenous canulation, use of new ProSeal LMA and care of donor site, etc. Patients who are severely neutropenic ($<0.5 \times 10^9$/L) should be nursed in isolation, receive prophylactic antibiotics and antifungals, regular mouth care including an antiseptic mouthwash such as chlorhexidine, and food of low bacterial content, as was done in our patient.[6] Laminar air flow facilities, although are not essential, should be used when available.

Adequate postoperative pain relief was rendered with IV tramadol to allow deep breathing exercises, adequate coughing and hence clearance of respiratory secretions to avoid pulmonary complications.

Long-term survival rates among children diagnosed with SAA are excellent due to the success of HLA-identical related hematopoietic stem cell transplantation, concurrent advances in immunosuppressive treatment and improved supportive care.[2] Perioperative management of aplastic anemia patients can have good outcome by preventing infection and efficient blood bank facility.

SUMMARIZING

1. Potential complications in aplastic anemia are caused by a decreased number or impaired function of all blood cells.
2. Standard sterile precautions along with smooth induction and emergence should be the anesthesiologist's goal.
3. A multidisciplinary team approach with coordination between the surgeon, hematologist, anesthesiologist and blood bank services can lead to a successful surgical outcome in such patients.

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